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## Case report

# Congenital subaortic stenosis by accessory mitral valve tissue, recognition and management

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## Abstract

Accessory mitral valve tissue as the single cause for left ventricular outflow tract obstruction is a very rare cardiac malformation in normally connected hearts. We report a case in which this condition was present as single cause for left ventricular outflow tract obstruction. The surgical technique is described and a review of the literature presented. © 1999 Elsevier Science B.V. All rights reserved.

**Keywords:** Subaortic; Stenosis; Accessory tissue; Surgery; Recognition

## 1. Introduction

Accessory mitral valve tissue causing left ventricular outflow tract obstruction is a rare pathologic finding in normally connected hearts. It is of great importance to repair the valve without valve replacement. A high incidence of reoperations have been reported in cases where it is not recognized or is incompletely removed [1]. When this pathology is associated with a classic fibro-muscular subaortic stenosis, it may remain unrecognized pre-operatively and result in an incomplete surgical resection. We report such a case and the tactics we employed to completely relieve the obstruction without causing valve dysfunction.

## 2. Case report

A 17-month-old child was referred due to progressive development of a left ventricular outflow tract obstruction. The diagnosis of subaortic stenosis had been established by echocardiography shortly after birth and the child was followed regularly. The child had a normal growth and never

presented signs of heart failure. On admission, the transthoracic echocardiography revealed a hypertrophied left ventricle and a severe subaortic stenosis with an estimated systolic gradient of 104 mmHg. No other cardiac anomaly was found. The analysis of the left ventricular outflow tract pointed out a subaortic ring-like membrane with a diameter of 9.7 mm and a normal mitral valve without a systolic anterior motion of the septal leaflet. The insertion of the mitral papillary muscles was normal. At that point, no abnormal valvular tissue was detected. Electrocardiography showed normal sinus rhythm and signs of right atrial overload. There were no clinical findings indicating congestive heart failure. The child was presented at our clinic for surgical treatment at 17 months of age, 9.4 kg in weight and 78 cm in height.

The child underwent surgery using cardiopulmonary bypass under moderate hypothermia. After aortic cross-clamping and delivery of cold-blood cardioplegia into the aortic root, the ascending aorta was opened. The aortic valve and the annulus appeared normal. A subaortic membrane was at the usual position and created a moderate stenosis. This membrane was resected en bloc from annular and subannular tissues using sharp and blunt dissection. After the resection the left ventricular outflow tract appeared to be wide open and the subvalvular mitral apparatus did not appear pathologic. Therefore, the aorta was

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closed, the left ventricle de-aired and the aortic clamp removed. Upon resumption of heart activity, the transesophageal echocardiography still revealed turbulent flow underneath the aortic valve. Direct measurement revealed a systolic pressure gradient of 70 mmHg between the left ventricle and the aorta, indicating a remaining stenosis. Cardiopulmonary bypass and aortic cross-clamping were reinstituted and a vertical left atriotomy was performed to inspect the mitral valve apparatus. A fibrotic mass attached to the mitral valve tissue was found close to the antero-lateral commissure. The tissue, including its subvalvular attachment was resected. Competence of the mitral valve was tested by injection of saline solution into the left ventricle. The left atrium was closed, the heart de-aired and the aortic clamp removed. Weaning from cardiopulmonary bypass occurred without difficulties. The TEE showed a non-turbulent flow between the left ventricle and the aorta and a competent mitral valve. Direct pressure measurement showed complete disappearance of the systolic gradient across the outflow tract.

The patients recovery was uneventful and she could be dismissed from the intensive care unit 2 days after surgery. After 11 months she remains in good general condition and shows normal development.

### 3. Discussion

To our knowledge, there are few such cases having been described until now. Usually there are coexisting cardiac malformations which may greatly influence the diagnostic pathway and the surgical approach. Most commonly this malformation is associated with ventricular septal defect (VSD) [2], transposition of great arteries (TGA) [3], coarctation, partial atrioventricular canal, double outlet right ventricle (DORV), membranous septal aneurysm (MSA) [4] and Noonan syndrome [5]. In this case, the accessory mitral valve leaflet was only associated with a small subaortic membrane.

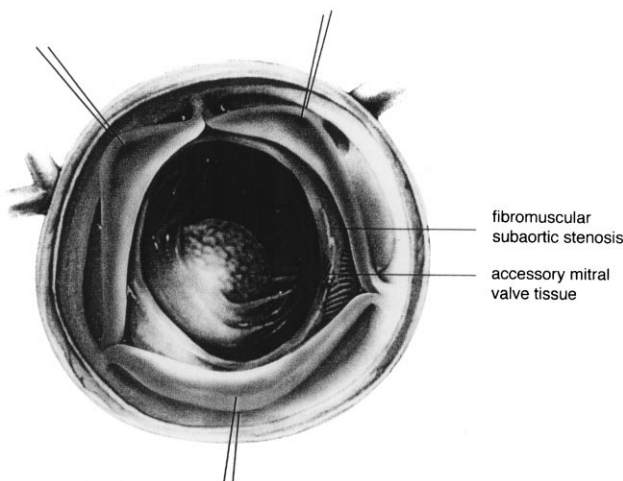


Fig. 1. Intraoperative transaortic view.

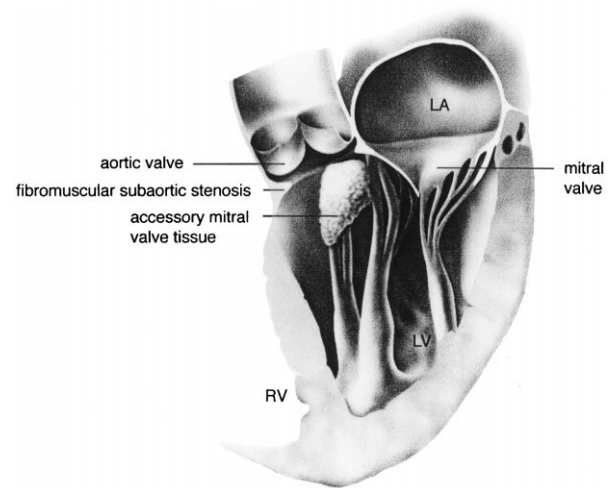


Fig. 2. Final appearance of fibrous obstruction as seen and resected through the mitral valve.

The correct preoperative diagnosis is extremely difficult to establish and is often ignored by conventional diagnostic methods. Echocardiography precisely delineates the left ventricular outflow tract obstruction and is frequently used as a sole diagnostic tool to assess subaortic stenosis [6,7]. Assessing the anatomic and dynamic contributions of the mitral valve in the obstructive process is also possible by echocardiography, although this analysis requires expertise and may be difficult to obtain preoperatively. Rare causes of left ventricular outflow tract obstruction arising from the mitral valve may remain unrecognized preoperatively, especially when a typical cause of obstruction, like a subaortic membrane is present. Detection of turbulent flow in post-repair transesophageal echocardiography pointed out the persistence of a significant outflow tract obstruction in this case. The fine analysis of the outflow tract revealed abnormal tissue in contact to the anterior mitral valve leaflet. The decision to approach this pathology through left atriotomy was made for two reasons.

One, remaining transvalvular pressure gradient after resection of the subaortic membrane with no abnormal tissue having been seen through the small aortic annulus after resection of this membrane, and two, competence of the mitral valve could be tested immediately and a repair procedure performed if necessary. The additional mitral valve tissue could not be properly identified at first, it appeared only as a mild valvular thickening (Fig. 1). The final pathologic appearance is shown in Fig. 2. Additionally, through left atriotomy the inspection of the mitral valve and its accessory tissue is easier, the resection of the accessory tissue, the corresponding chordae and papillary muscle can be accomplished with great precision and without possibly damaging other structures. Ascutito et al. [8] describe a similar case in which the initial transaortic approach was unsuccessful and left atriotomy had to be performed. Yasui et al. [2] describe the approach through left atriotomy, but their cases all had coexisting cardiac malformations which

enabled them to approach through an enlarged ventricle septal defect. Sono et al. [3] report a case where initial aortotomy followed by left ventriculotomy was used and Meldrum et al. [9] report a case where transaortic approach was successful. Szedo et al. [1] report a case where transaortic approach was successful upon reoperation. These cases stress the importance of controlling the relief of obstruction even in apparently straightforward cases. In this regard direct measurement of the gradient and operative TEE are most important in assessing the residual stenosis. TEE, furthermore, provides an anatomic definition of the outflow tract and may point out rare anomalies. A second look through left atriotomy appears advantageous because a wider view of the mitral subvalvular apparatus is obtained, allowing precise excision of the abnormal tissue. Although it requires expertise, direct cardioscopy through the aortic annulus may provide an alternative to this approach.

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